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**Pseudoxanthoma Elasticum and Clotting Factor Deficiency in a Pregnant Patient**

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**Introduction**: Pseudoxanthoma elasticum (PXE) is a connective tissue disease that causes skin laxity in the neck, axilla, groin, and flexural creases. Retinal hemorrhages are common. PXE can be associated with deficiency of vitamin K-dependent clotting factors. There are two variants of this disease: an autosomal recessive condition that presents in infancy and is usually fatal, and a milder form, probably autosomal dominant, that usually presents in female adolescents.

**Case**: A 19 yo G1P0 woman with PXE presented for care at 30 weeks gestation. She was diagnosed with PXE at age 13 when she developed excessive skin folds. The diagnosis was confirmed by skin biopsy. Her younger sister had a more involved course, with cardiac and renal involvement. A maternal aunt had mild disease.

A deficiency of vitamin K-dependent clotting factors was diagnosed. Lab values showed decreased activity in Factors II (43%), VII (31%), IX (56%), and X (18%). Her INR was elevated at 1.92, and PTT remained normal at 32 seconds. While the patient noticed occasional bruising, she had no significant epistaxis, menometrorrhagia, or gingival bleeding. High-dose oral vitamin K failed to correct her clotting factors. Genetic testing identified a mutation in the GGCX gene, which controls the production of vitamin K-dependent clotting factors. Options for correcting this clotting factor deficiency included FFP and/or factor IX concentrate (Bebulin®). Factor IX concentrate is used to treat minor bleeding and is roughly equivalent to 3 units of FFP.

However, when the patient presented to the labor and delivery floor at 36 6/7 weeks gestation, her obstetrician and hematologist opted not to correct her clotting factor deficiency. Therefore, we provided intravenous PCA (PCIA) analgesia with ketamine 1mg/mL and fentanyl 10 mcg/mL at a dose of 1 mL q 10 min. We started without a basal infusion, but then increased the basal infusion to 1 and then 2 mL/hour. Although the patient was nulliparous, her labor was short; she delivered 5 hours after the PCA infusion began. The patient had no postpartum complications, and her neonate had no respiratory depression.

**Discussion**: Providing adequate analgesia to patients with contraindications to neuraxial analgesia is difficult. PCIA fentanyl or remifentanil are frequently used. However, fentanyl is associated with neonatal depression and maternal sedation, nausea, and poor pain relief. Remifentanil is only approved for use with an anesthesia provider in constant attendance, which is labor-intensive. Ketamine, an NMDA-receptor antagonist, prevents fentanyl-induced hyperalgesia and permits lower fentanyl doses, which enhances safety. Ketamine improves the analgesia from PCIA fentanyl post-operatively but the use of this combination in labor has not been reported.

**References**  